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Polioencephalitis Superior (Nuclear Ophthalmoplegia) and Poliomyelitis.

BY

B. SACHS, M. D.,

Professor of Mental and Nervous Diseases in the New York Polyclinic.

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POLIOENCEPHALITIS SUPERIOR (NUCLEAR OPHTHALMOPLEGIA) AND POLIOMYELITIS.[1]

By B. Sachs, M.D.,
PROFESSOR OF MENTAL AND NERVOUS DISEASES IN THE NEW YORK POLYCLINIC.

It is a rare experience in neurological matters to have the pathology of a disease unravelled as quickly as was done in the case of those clinical groups of symptoms which we know as ophthalmoplegia externa and interna.

The paralysis of the muscular apparatus of the eye was soon discovered to be due, in most cases, to a lesion or lesions affecting the nuclei of the nerves which govern the various ocular muscles. In this nuclear paralysis, the nuclei of the oculo-motor nerves play a most important *rôle*, though the nuclei of the fourth and sixth nerves are involved frequently enough. Our knowledge of the pathological processes resulting in such lesions was gained by showing first of all that all the cranial nerve nuclei were subject to the same acute and chronic affections, and that what we ordinarily call progressive bulbar paralysis was, as far as its pathological character is concerned, exactly the same as the progressive paralysis affecting the ocular nuclei.

The relation of the bulbar process to poliomyelitis was firmly proved by cases of bulbar paralysis which were associated with symptoms resembling those of a progressive muscular atrophy or a chronic anterior poliomyelitis, and furthermore by cases of typical progressive muscular atrophy which, in their terminal stages, developed bulbar symptoms.

On the strength of this clinical analogy, Hutchinson, Mauthner, and Birdsall were struck with the pathological resemblances between the diseases affecting the ocular and spinal nuclei; and Wernicke proposed to call the affection of the oculo-motor nuclei a polioencephalitis superior, whence it followed that the bulbar paralysis might well be styled polioencephalitis inferior. While the analogy with poliomyelitis had been proven beyond the shadow of doubt for the bulbar cases, Wernicke's theory needed further proof as regards the cases of total ophthalmoplegia externa and interna. Heretofore but one case has been recorded (by Seeligmüller) in which the symptoms of a chronic

[1] Read before the American Neurological Association, June, 1889.

poliomyelitis were associated with those of polioencephalitis superior, and with the exception of the cases of Henoch and Buzzard[1] in which an ocular (nuclear) paralysis occurred in the course of an acute poliomyelitis anterior, I know of no cases which exhibit this interesting association of symptoms. The case which I report here proves rather conclusively that the same pathological process may give rise to a typical polioencephalitis superior and to chronic or subacute poliomyelitis anterior.

The history of this case is as follows:

H. M., æt. forty, is a man of robust build, unusually intelligent, and one who has experienced all the vicissitudes of life. He was born in this city, and has been married fourteen years. Has one brother living and healthy, and one brother who died from want of water, as the patient says, on the desert of Arizona. Both parents are dead; the father died of yellow fever in New Orleans in 1858; the mother died of a paralysis which lasted seven or eight years and began by turning-in of both feet. The paralysis of the legs became complete; she died at the age of seventy-four. The patient went to school at Syracuse, N. Y. As a boy had frequent "bilious" attacks associated with headaches and vomiting, was otherwise in robust health. He attended school until the age of twelve; went to New Mexico at the age of fourteen, and there learned a trade.

When sixteen years of age, one day after reading several hours got up to stretch himself, but fell back unconscious against a hot stove, burning the left temple severely—the scar is visible at the present day. He was insensible for several hours and was then put to bed; knew nothing of what had happened until he saw doctors around; no paralysis followed. A second similar attack, again after reading, occurred three months later, from which he quickly recovered. A third attack occurred, but the date of this he cannot recall. No further sickness until the age of twenty, when he was in Peru and was steward on a United States steamer. While on shore he fell from a horse, striking the right elbow and injuring the arm. Recovered completely from this fall.

At the age of twenty-five was in Europe; felt one day a severe pain in the left eye; engaged passage at once for New York, reaching there a few weeks later. By that time the left eye was closed. Went to sea again (to Australia on a sailing vessel), and during this trip noticed that the right lid was also beginning to droop. He was treated in Melbourne by electricity, but the condition remained stationary for several months. After that a slight improvement is said to have set in in the left eye, but the right eye grew worse; both pupils were dilated (physician's statement). Had double vision all the time, and small ulcers formed on the left eye. The Australian physicians suspected tumor of the brain. He had severe headaches at the time; the left eye became inflamed and was in such a bad condition that the physician advised enucleation, but the patient objected. After a short trip at sea, the inflammatory condition was at an end.

It was about this time that he began to be suspicious of his legs, for

[1] Quoted by E. Blanc, Arch. gén. de Méd., Janv. 1887.

one day while walking on the deck his right knee gave way. A few days later the same accident occurred. Nevertheless he joined a ship from Australia to California. When thirty days out, he had to refuse duty as steward, as he could not move his right thigh, leg, or toe a single inch. The captain ordered hot steam bath, and gave him blue pill and black draught. He went to Oregon next—now thirteen years ago. He remembers that when there he could not hold water nor contain feces. His left leg was not affected at any time. Iodide treatment was proposed; patient objected on the ground that he had never had any syphilitic affection. In consequence of the paralysis of the right leg he was compelled to go about on crutches. The doctor who examined him found a sensitive point between the shoulder-blades and ordered blisters, and strychnine internally. In six months time he was able to walk with the assistance of a stick; he could use the hip and knee joint, but could not move ankle or toes. The eyes remained in about the same condition; he was not worried by them. He undertook contract work on a railroad in Panama, and there contracted severe malaria.

One year and a half ago he was stricken down with prolonged fever. As soon as he recovered from this he travelled about considerably; finally went to Jacksonville, where he got a thorough drenching, which was followed by severe chill. One day he tried to read the papers but could not see anything. Last year, on his return to New York he was examined by Dr. E. Grüning, who performed an iridectomy of the right eye which did not improve vision. Later on, Dr. Grüning raised the left eyelid by operation and restored vision to that eye. Went to Panama in May, 1888, and returned to New York about seven weeks ago.

For the past four weeks he has been an inmate of the Montefiore Home for Chronic Invalids, where I have had the opportunity of studying the case. He denies ever having had gonorrhœa or syphilis, and an examination of his body reveals no symptoms of the latter. Has been moderate in sexual matters and has never been a hard drinker. Has smoked innumerable cigarettes for years. No loss of consciousness has occurred since the attacks recorded above. Does not suffer from headaches and but for the condition of his eyes and of his right leg would feel entirely well.

Present condition.—Strong, well-built man; heart sounds normal; no enlargement of liver, slight enlargement of spleen; other thoracic and abdominal organs normal. The most striking feature of the patient's appearance is the double ptosis, at present more marked on the right than on the left, in consequence of the operation on the left eyelid. Slight lateral nystagmus of right eye; in this eye also maculæ corneæ, old iritis with exclusion of pupil, small coloboma upward. Right pupil is not visible, left pupil dilated. A transparent, thin membrane has grown upward, covering nearly one-half of left pupil. The results of my examination of the eyes, which were kindly corroborated by Dr. E. Fridenberg, are as follows:

O. D.—Paretic—rectus internus, rectus externus, and inferior oblique. Paralyzed—levator palpebrarum, rectus superior, obliquus superior, and rectus inferior. Associated movements with the left eye do not differ from those attempted singly.

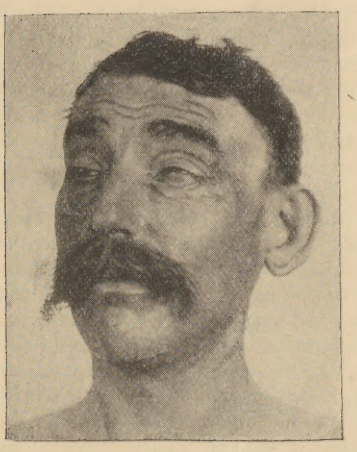
O. S.—Paretic—rectus externus, rectus internus, and superior oblique. Paralyzed—levator palpebrarum, rectus superior, rectus inferior, and obliquus inferior. Paralysis of iris (light reflex abolished), ciliary mus-

cle normal. Accommodation reflex good. Media apparently clear in both eyes. Vision, left eye, $\frac{20}{C}$. Right eye = 0.

Ophthalmoscopic examination.—Left papilla normal; right papilla cannot be examined.

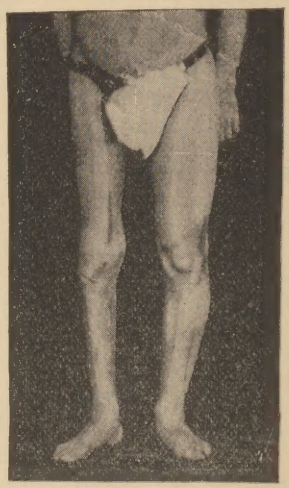
No change in the facial distribution. Hearing normal on right side; on left side somewhat diminished, but normal bone conduction. Tongue protruded straight, slight fibrillary movements. Sensation of face and tongue normal in every particular. Smell and taste normal on both sides. The left arm appears to be slightly larger than the right, but grasp is equally strong on both sides. Sensation normal to touch and pain. Distinguishes numbers written on arm with greatest ease. No reflexes to be obtained in upper extremities. No difficulty in respiratory or abdominal muscles.

FIG. 1.



Subject attempting to look to extreme right.

FIG. 2.



Showing atrophy of right leg.

Lower extremities.—Marked atrophy of right leg from hip downward. Largest circumference on right side; hip, 14¼ inches; left side, 19½ inches; right calf, 10 inches; left calf, 12½ inches. Patient can flex knee very little, but cannot move toes of the right foot. Walks by excessive use of posterior thigh and leg muscles. Muscular excitability lost in right thigh. No disturbance in sensation except apparent diminution of pain sense on inner aspect of right thigh. The left thigh and leg muscles show normal myotatic excitability and absolutely normal sensation. No change to be noted in any respect in leg of left side. There is no ataxia of either leg and none in the upper extremities. No Romberg symptom. The knee-jerks are lost on both sides and cannot be elicited by Jendrássik's method. All cutaneous reflexes sluggish but present.

The electrical examination reveals no changes in any of the muscles

of the face, of the upper extremities, or of the trunk, nor in the left leg, but marked degeneration reaction exists in the anterior thigh and leg muscles. The vasti and anterior tibial muscles are atrophied to such an extreme degree that no reaction could be obtained with currents at command.

This history can be summarized in a few words: A man in perfect health, without any specific alcoholic or hereditary taint, is affected with a slowly developing paresis or paralysis of all of the ocular muscles. This condition is scarcely fully established before a weakness of the right leg is noticed by giving way of the knee. This weakness is developed into a most marked paralysis associated with extreme atrophy. The symptoms remain restricted to the right leg, become retrogressive, and have not to this day affected the opposite leg. The arms remain entirely normal. The transitory bladder and rectal symptoms were probably due to an extension of the inflammation of the gray matter, and do not imply, to my mind, the existence of a transverse myelitis, acute, subacute, or chronic. No other interpretation can be put upon these symptoms except to say that in the course of a chronic nuclear paralysis of the eyes a subacute poliomyelitis set in. Both in the eyes and in the leg the disease developed in the same fashion and has practically remained stationary for years.

It will hardly be necessary in this paper to prove the diagnosis of subacute poliomyelitis in this case, and considering the rarity of poliomyelitis in the adult it would be strange indeed if the occurrence of such an affection in the course of a polioencephalitis superior were a mere coincidence. It seems to me to prove positively that the ganglion cells of the anterior horns of the spinal cord are subject to the same pathological changes as the large nuclear cells on the floor of the third and fourth ventricles.

In the case which Seeligmüller described, the upper and lower extremities were the parts first to be affected, and secondarily an affection of the eye, right and left oculo-motor nerves, set in. In this case there would seem to be a spreading by contiguity; and yet the third, fourth, and sixth nuclei were affected before the seventh, tenth, eleventh, and twelfth nuclei. The absence of such contiguity in my case does not argue, I think, against the supposition that the leg and eye symptoms are dependent on the same pathological process. It is a peculiarity of this morbid process that it is selective, that in some cases it affects parts contiguous with one another, but that again in other cases it does not spread in this manner. In some instances the upper extremities are affected and with these the ocular muscles, showing that the bulbar nuclei have not been similarly affected. The bulbar nuclei and oculomotor nuclei may be affected without an involvement of the sixth and seventh nuclei, and among the various cell groups constituting the

oculo-motor nuclei we find that the pathological process may attack portions of the nuclei which are situated at the extreme anterior and posterior limits of the nucleus and skip cell groups lying in between. This is in close keeping with the pathological condition we meet with in cases of poliomyelitis anterior, for we see too many of these cases not to know that contiguous muscles often escape paralysis and atrophy, simply because their representative ganglion cells in the spinal cord have not been affected like their immediate neighbors.

The involvement of the iris in the one eye (the condition of the other could not be examined) takes my case out of the category of cases of ophthalmoplegia externa. According to most authors, an ophthalmoplegia externa, with paralysis of the iris, would compel one to refer the lesion to the base of the brain, but since Westphal and Spitzka have plausibly shown that the nuclei for the accommodation and light reflex lie anteriorly and away from the remaining oculo-motor nuclei, it is readily seen that these nuclei also, one or both, may be affected by the extension of the inflammatory process. It is in this way that I explain the affection of the iris in this case. Since the accommodation reflex remained normal, it is natural to infer that the ciliary and iris nuclei must be some appreciable distance apart. We must be careful, however, not to be too positive in such assertions, for Thomsen has recorded cases in which there were distinct paralyses of various ocular muscles with only the slightest involvement of a few of the nuclear cells, and, strangest of all, one case of paralysis of associated vision upward due to a gummatous infiltration of the *oculo-motor root fibres*, whereas the nuclei were found entirely normal. It is for this reason also that I believe that the determination of the exact location of the various subdivisions of the oculo-motor nucleus on clinical grounds only,[1] has been carried too far. This question can be settled in no other way but by the experimental method, or by noting to what extent clinical and post-mortem records tally.

One other point in the case demands explanation: the knee-jerk is absent on both sides. The first suspicion was that of an accompanying tabes dorsalis, as in Westphal's well-known case; but this supposition must be abandoned, since a close examination with this end in view has shown the absence of every other important symptom of tabes. The absent reflex on the left side must therefore be regarded as the only evidence of the extension of the process in the spinal cord to the left half of the cord, but at the same time the normal condition of the muscles, the normal electrical reactions, and the total absence of atrophy prove that that side can be affected but very little.

The chief value of my case is, that it proves the close relationship

[1] Starr, Journal of Nervous and Mental Diseases, May, 1888.

between the gray matter at the floor of the third and fourth ventricles and the anterior gray horns of the spinal cord.

Wernicke choose the term polioencephalitis superior wisely enough; but Strümpell's polioencephalitis, a supposed cortical disease, has caused some confusion. Strümpell's theory and disease lack proof, and for the present we need not decide whether we shall have to add a polioencephalitis suprema to polioencephalitis superior.

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